

CASE REPORT

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Surgical Treatment of Cavernous Malformation of Pineal Region

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The main causes of pineal apoplexy are hemorrhage associated with pineal region tumors, vascular malformations, and pineal cysts. Cavernous malformations rarely occur in the pineal region, with only fifteen cases reported previously. Hemorrhage associated with cavernous malformation causes apoplectic event in the pineal region. We report two surgically treated cases of pineal hemorrhage associated with cavernous malformation and discuss the consideration in management of the pineal apoplexy.

KEY WORDS : Pineal apoplexy · Hemorrhage · Cavernous malformation.

Introduction

Pineal apoplexy is one of pineal region syndrome secondary to hemorrhage in pineal region^{1,8,9)}. The causes of hemorrhage in pineal region are associated with pineal cyst, pineal tumor or adjacent vascular malformation^{1-3,8,9,14)}. We present two cases of pineal apoplexy with hemorrhage in pineal cavernous malformation treated with resection. Up to date, only one report presented successful conservative operative management of pineal apoplexy with ventricular shunting to relieve acute obstructive hydrocephalus and other reports advocated resection of the pineal region^{2,9,11)}. Therefore we discuss the strategies for managements of pineal apoplexy.

Case Report

Case 1

A 36-year-old man complaining of headache, diplopia, and posterior neck pain was referred to our hospital. His brain computed tomography(CT) scan showed mass with spotted calcification and acute hemorrhage in pineal gland area and third ventricle (Fig. 1A). This hemorrhage showed mixed signals of different ages, but no hemosiderin rim on magnetic resonance image (Fig. 1B, C). We thought that the cause of hemorrhage was due to tumor such as germ cell tumor or pineal cyst. We were easily able to rule out the possibility of vascular

malformation because magnetic resonance angiography revealed no vascular abnormalities. Serial brain CT scans which performed weekly after the first attack of symptoms showed resolving hemorrhage and progressively enlarged ventricle (Fig. 1D). After we treated him with conservative care for two weeks, he experienced an improvement in diplopia, but his headache become progressively more severe. We performed ventriculoperitoneal shunting using Codman Hakim programmable valve shunt (Codman & Shurtleff Inc/Johnson & Johnson Co, Raynham, MA) system. Two days after shunting, he complained of a sudden aggravation of his diplopia. Follow-up brain CT scan showed rebleeding from the previous lesion (Fig. 1E). The mass and the hematoma were removed totally via occipital transtentorial approach. The dark red mass had a thin capsule and easily separated from the surrounding tissues. Most of the mass consisted of old hematoma and there was no apparent tumor tissue. These surgical findings corresponded to a cavernous malformation. On histologic examination, there were large hematoma and many vascular spaces which were lined by a single layer endothelium. The diagnosis was pineal apoplexy with cavernous malformation. He was discharged without neurological deficit.

Case 2

A 42-year-old man visited our hospital due to severe headache and suddenly developed diplopia. On neurological

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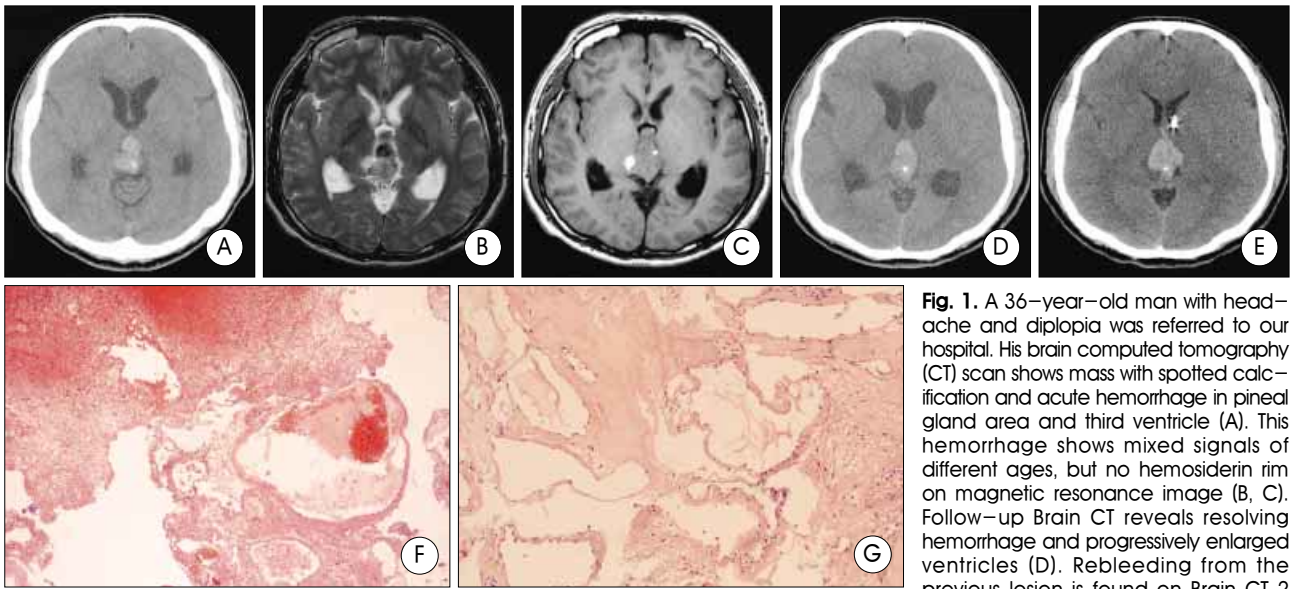


Fig. 1. A 36-year-old man with headache and diplopia was referred to our hospital. His brain computed tomography (CT) scan shows mass with spotted calcification and acute hemorrhage in pineal gland area and third ventricle (A). This hemorrhage shows mixed signals of different ages, but no hemosiderin rim on magnetic resonance image (B, C). Follow-up Brain CT reveals resolving hemorrhage and progressively enlarged ventricles (D). Rebleeding from the previous lesion is found on Brain CT 2

days after shunting (E). The mass and the hematoma were removed totally via occipital transtentorial approach. The dark red mass had a thin capsule and easily separated from the surrounding tissues. Most of the mass consisted of old hematoma and there was no apparent tumor tissue. These surgical findings corresponded to a cavernous malformation. On histologic examination, there were large hematoma and many vascular spaces which were lined by a single layer endothelium. Hematoma and variable sized vascular channel are noted (H&E, x40) (F). Hyalinized vascular channels without intervening glial tissue are noted (H&E, x100) (G).

examination, he had a limitation in upward gazing of both eyeballs without papilledema and his mental status was nearly alert. Magnetic resonance imaging and brain CT scan showed acute hemorrhage and abnormal mass in the pineal gland area (Fig. 2A, B). We should first consider the cause of pineal hemorrhage.

Tumors, vascular malformations, and cysts were thought to be the cause of this lesion. 4-vessel cerebral angiography and the study of serum tumor markers such as betahuman chorionic gonadotropin, alpha-fetoprotein, and placental alkaline phosphatase were performed. But there was no vascular anomaly and abnormal tumor markers. Patient's condition progressively deteriorated during our evaluations for differential diagnosis.

The presence of a newly developed hemorrhage and hydrocephalus was noticed at his follow-up brain CT scan (Fig. 2C). For the management of acute hydrocephalus, Ventriculo-peritoneal shunting with a Heyer-Schulte high-pressure valve shunt (Heyer-Schulte Co, Goleta, CA) system was performed (Fig. 2D). But the patient's condition became worse and mental status was stuporous 2 weeks after shunting. Second follow-up brain CT scan showed newly developed huge hemorrhage and aggravated hydrocephalus (Fig. 2E). The mass and the hemorrhage were totally removed via an occipital transtentorial approach. The mass was filled with xanthochromic, reddish brown colored old blood, and mixed with acute hematoma. The patient recovered his consciousness with some limitation of upward gaze remaining. Histologic examination revealed the

presence of a cavernous malformation composed of a compact mass of sinusoidal-type vessels.

Discussion

Pineal apoplexy is a very rare event, with only 12 cases reported previously^{1-3,9}. Apoplexy means ill-defined symptoms consequent on acute hemorrhage in a normal structure or lesion². Therefore, pineal apoplexy is various symptoms related with the acute hemorrhage originating in a pineal cyst, or pineal region tumors, or adjacent vascular malformation^{1-3,9,11,14}. But there has been no report of pineal apoplexy into a normal gland^{2,9}.

We experienced two case of pineal apoplexy subsequent to acute hemorrhage into a pineal cavernous malformation. Pineal apoplexy related with cavernous malformation is very rare. Since cavernous malformation of pineal region was first described by Miller in 1961¹, only fifteen more cases have been reported^{4,6,7,9,12,13,15,16}. These lesions account for less than 1% of cavernous malformations of central nervous system¹⁷. The hemorrhage of pineal region compress the posterior thalamus and upper brain stem in consideration of the anatomical location of pineal region¹⁶. Moreover, once hemorrhage had occurred into cavernous malformation, the rebleeding rate is relatively high⁸. In present cases, two patients all experienced repeated hemorrhage at least three times until the surgical resection of pineal region and hematoma was performed.

Up to date, only one report presented case of successful ma-

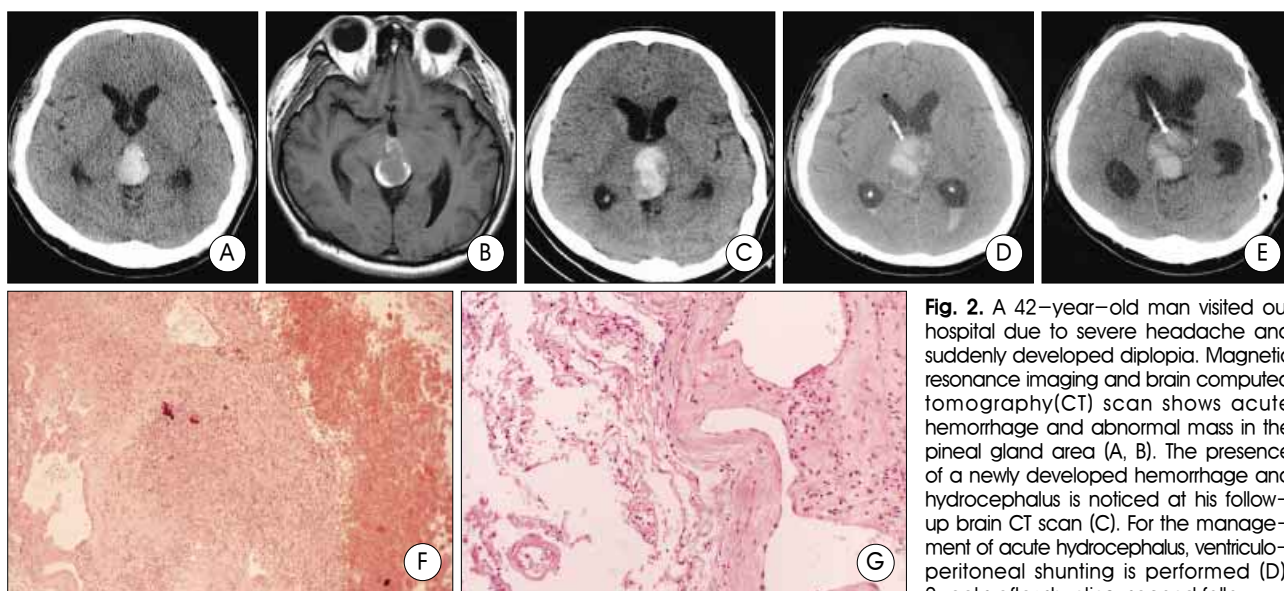


Fig. 2. A 42-year-old man visited our hospital due to severe headache and suddenly developed diplopia. Magnetic resonance imaging and brain computed tomography(CT) scan shows acute hemorrhage and abnormal mass in the pineal gland area (A, B). The presence of a newly developed hemorrhage and hydrocephalus is noticed at his follow-up brain CT scan (C). For the management of acute hydrocephalus, ventriculo-peritoneal shunting is performed (D). 2 weeks after shunting, second follow-up

brain CT scan shows newly developed huge hemorrhage and aggravated hydrocephalus (E). The mass and the hemorrhage were totally removed via an occipital transtentorial approach. The mass was filled with xanthochromic, reddish brown colored old blood, mixed with acute hematoma. Histologic examination revealed the presence of a cavernous malformation composed of a compact mass of sinusoidal-type vessels, hematoma (right side) and aggregation of hemosiderin laden macrophages are seen (H&E, x40) (F). Variable sized hyalinized vascular channels are noted (H&E, x100) (G).

management of pineal apoplexy by conservative operation that is ventriculo-peritoneal shunt without resection of lesion²⁾. Most other authors have recommended surgical removal of apoplectic pineal lesion as either a primary treatment or following operative cerebrospinal fluid diversion for the risk of repeated hemorrhage and progressive brain stem dysfunction^{2,5,9,11,18)}.

Although there is one case that conservative operative management was successful in the treatment of pineal apoplexy, surgical resection is the treatment of choice of pineal apoplexy regardless of the causes because of repeated bleeding and progressive neurological deterioration^{2,5,9,11,18)}. In the case of successful conservative operative management of pineal apoplexy, the patient had been treated with oral anticoagulation for pulmonary thromboembolic disease and had head trauma history before a symptom developed. Also she was too old to undergo surgical resection of pineal lesion due to the risk of mortality and morbidity. However, shunting before the resection of pineal cavernous malformation could have been very dangerous, because it might have caused huge hemorrhage originating into the lesions, which could have caused a sudden deterioration in the patients condition. Therefore, we recommend that spontaneous pineal apoplexy without underlying bleeding tendency should be treated with surgical extirpation of pineal region to prevent the repeated hemorrhage and the progressive neurological deterioration due to mass effect of repeated hemorrhage to the posterior thalamus and the upper brain stem.

Conclusion

Although pineal apoplexy have several different causes, such as pineal cyst, pineal tumor or adjacent vascular malformation, spontaneous pineal apoplexy without underlying bleeding tendency should be treated with surgical resection of pineal lesion regardless of the causes to prevent repeated hemorrhage and progressive neurological deterioration.

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